A rare case of giant congenital melanocytic nevus associated with ipsilateral developmental dysplasia of the hip

Ipsilateral gelişimsel kalça displazisinin eşlik ettiği dev konjenital melanositik nevüsü nadir bir olgu

Meryem Aktaş, Deniz Yücelten, Özlem Akın Çakıcı, Evrim Karadağ Saygı

Marmara University Faculty of Medicine, Department of Dermatology; *Department of Physical Therapy and Rehabilitation, Istanbul, Türkiye

Keywords: Congenital, melanocytic nevus, developmental hip dysplasia

To Editor,

Congenital melanocytic nevi (CMN) are benign melanocytic proliferations that typically present at birth. Giant congenital melanocytic nevi [(GCMN), estimated adult size >40 cm] is of great importance due to its association with serious complications such as malignant transformation and/or central nervous system involvement in addition to psychological burden, itching, and many accompanying disorders impairing quality of life for patients and their families.

Here, we aim to highlight a GCMN case with developmental dysplasia of the hip (DDH), where limited data are available in the literature. A 5-month-old girl was referred to our department for having a GCMN. She was born to a 28-year-old mother in the 33rd week of gestation after a pre-eclamptic pregnancy. Her parents had no consanguinity or family history of DDH. During routine examination in the fourth week of infancy, hemihypertrophy and asymmetry of the gluteal folds, leg length difference, and extension limitation in the right knee had been detected. However, the acetabulum and femoral head relationship was reported as within normal range for both hips by ultrasonography. Patient informed consent was obtained from her mother as the patient’s guardian.

Cutaneous examination revealed a darkly pigmented CMN occupying the left gluteal area along with the right lumbar and lower abdominal area and right lower extremity excluding the forefoot (Figure 1A, B). There were 11 satellite lesions scattered on the left hand and leg, the upper part of the trunk, and the face. Her right thigh was hypertrophic, and her right leg was approximately 5 cm shorter, besides 30 degrees of passive and active extension limitation. Otherwise, she was healthy. A magnetic resonance imaging was performed to evaluate for neurocutaneous melanocytosis (NCM), and it was normal.

DDH was diagnosed by detecting dislocation in the right femoral head on control pelvic plain radiography at the age of 1 year (Figure 2). At the age of about 2 years, the patient had a history of fracture of the right femur as a result of falling, but the bone union was completed. She used an adjustable knee orthosis for about 3 years. Extension limitation in the right knee regressed with orthoses, but leg length discrepancy persisted. Orthopedic intervention was performed at 4 years of age (a delay owing to the coronavirus disease-2019 pandemic) as a result of insufficient response to conservative treatments. No suspicious change was detected in GCMN during 4.5 years of clinical follow-up.

Cite this article as: Aktaş M, Yücelten D, Akın Çakıcı Ö, Karadağ Saygı E. A rare case of giant congenital melanocytic nevus associated with ipsilateral developmental dysplasia of the hip. Turkderm-Turk Arch Dermatol Venereol. 2024;58:52-3

ORCID: orcid.org/0000-0002-0157-0748

Copyright © 2024 The Author. Published by Galenos Publishing House on behalf of the Society of Dermatology and Venereology. This is an open access article under the Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 (CC BY-NC-ND) International License.
DDH is a spectrum of clinical and imaging findings ranging from mild instability to overt dislocation with an incidence of 1/100-1000. In the pathogenesis, the instability between the femoral head and acetabulum initiates the dysplasia process and hypertrophy of ligaments, cartilages, pulvinar (fibrofatty tissue in the joint) in subsequent stages causes progressive deterioration of joint anatomy and hip dislocation may occur.

Soft tissue hypertrophy/hypotrophy, lipomatosis, and spina bifida have been associated with CMN along with well-known complications such as NCM and malign melanoma. However, there is not enough data to establish a definite causality. Many local mediators such as platelet-derived growth factor, transforming growth factor-alpha (TGF-alpha), TGF-beta (β), and interleukin-1 have been shown to increase in melanocytic nevi. Disequilibrium in the level of these mediators that play an important role in the support and synchronization of growth and development may cause disorders of other structures developing in the nevus area. Accordingly, data are showing that the TGF-β polymorphism may be a risk factor for DDH.

Although it has been reported that genetic factors may be involved in the pathogenesis of DDH in addition to environmental factors, it is hard to associate DDH with CMN on genetic basis to date. Given the available data and the pathogenesis of DDH, it seems more reasonable to consider the relationship between DDH and CMN as a disruption of immunohistochemical and anatomical synchronization during development.

Finally, GCMN affecting pelvic girdles may be a risk factor for DDH, however more extensive studies are warranted to reveal this association more precisely and to define the predictive factors.

**Ethics**

**Informed Consent:** Patient informed consent was obtained from her mother as the patient’s guardian.

**Authorship Contributions**


**Conflict of Interest:** No conflict of interest was declared by the authors.

**Financial Disclosure:** The authors declared that this study received no financial support.

**References**